

Sjögren's Syndrome

- Epidemiology
 - Adult female
- Mechanism
 - dysfunction of exocrine glands due to lymphoplasmacytic infiltration
 - can be primary or secondary (RA, Scleroderma, SLE, PM)
- S/S
 - $\geq 4/5$ Diagnostic Criteria (94% sensitive and 95% specific)
 - **(1) Dry Eyes aka Xerophthalmia** (never ask about dry eyes rather ask if pt has the sensation of foreign body, gravel, burning, light sensitivity, excessive mucuous accumulation in the morning, et al)
 - **(2) Schimer Test** (filter paper is placed at the palpebral fissure and after 5min if $<8\text{mm}$ of the paper is wet then positive) or **Rose-Bengal Test** (fluorescein dye is placed onto eye and if there is devitalized/damaged conjunctival epithelium then positive)
 - **(3) Dry Mouth aka Xerostomia** (never ask about dry mouth rather ask if pt drinks water in the middle of the night, constantly chews gum, has difficulty chewing/swallowing, has difficulty talking, et al)
 - **(4) Inner Lip Biopsy** (if lymphoplasmacytic infiltrate in minor salivary glands then positive)
 - **(5) Auto-Ro/SS-A** (55% sensitive) or **Anti-La/SS-B** (40% sensitive) SS = Sjogren's Syndrome
 - Other S&S
 - PEx: No Salivary Pool in Vestibule, Dry/Red/Smooth Tongue, Parotid Gland Enlargement (always exclude infection, cancer, infiltrative disorders below, bulimia), Cheilitis, Gingivitis, Poor Dentition
 - Abs: ANA (95% sensitive) RF (75% sensitive)
 - Cancer: **44x increased risk of MALToma and Waldenstrom's Macroglobulinemia**, disappearance of RF is a marker, always check for LAD and splenomegaly
 - General: Extreme Fatigue, Photosensitivity
 - MS: Symmetric Polyarthritis of the Hands, Myalgias
 - CV: Pericardial Effusions, Vasculitis, Raynaud's
 - Pulm: URT Infections esp Sinusitis, ILD, Pleuritis, Dry Cough
 - GI: Pancreatitis, Biliary/Liver Disease b/c of the high association with PBC/AIH
 - GU: Vaginal Dryness
 - Renal: Type I RTA, Nephrogenic DI
 - CNS: Various Neuropathies, Psych Problems
 - Endo: HypoTH
 - Derm: dry skin but also erythematous nodules/plaques/patches
- DDx
 - Age Related Sicca
 - Anticholinergics
 - Diabetes can give you dry mouth and blurry vision 2/2 hyperglycemia
 - Infiltration (Lymphoma, Sarcoid, Hemochromatosis, Amyloid, Hyperlipidemia)
 - Infection (HIV, HBV, HCV, EBV, TB, Syphilis)
- Tx
 - Eye
 - Mild: Artificial Tears (NOT Vizine), avoid agents that reduce lacrimal flow (diuretics, antidepressants, antihypertensives, et al)
 - Mod: Tear Duct Plugging / Electrocauterization
 - Severe: Immunosuppressants w/ ophthalmic cyclosporine (Restasis)
 - Mouth
 - Mild: Sugar Free Gum/Candy (nothing with sugar b/c pts have bad dental problems), Drink Lots of Water, Good Dental Care, Antifungals b/c pts often get thrush b/c they are on steroids
 - Mod: M3 Cholinergic Agonist w/ cevimeline (Evxac) or pilocarpine (Salagen)
 - Systemic
 - Mild: NSAIDs
 - Mod: Steroids
 - Severe: first hydroxychloroquine (Plaquenil) then methotrexate